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Case Report

Primary Kidney Lymphoma Mimics Renal Cell Carcinoma in Preoperative Liver Transplant Patient: Postoperative Challenge – A Case Report

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Abstract

Cancer screening is a critical component of the pretransplantation process. However, in patients with deteriorating liver function, the focus may shift towards acquiring a functioning liver, potentially delaying cancer evaluation.

A 42-year-old male with primary sclerosing cholangitis (PSC), CTP score C10, and MELD score 19 was evaluated for liver transplantation (LT). Preoperative imaging revealed two lesions in the upper pole of the left kidney, raising concern for malignancy. After discussion at a uro-oncology meeting, the decision was made to resect the lesions post-LT. On postoperative day 5, the patient underwent entero-enterostomy revision and resection of the renal lesions due to melena. Pathological analysis confirmed Burkitt lymphoma/high-grade B-cell lymphoma-NOS. The patient completed four cycles of chemotherapy and remained free of recurrence by the second postoperative year.

This case underscores the importance of cancer screening in LT candidates and highlights the need for biopsy when radiologic suspicion arises. Management should be individualized, considering both liver disease severity and malignancy, with a tailored treatment approach.

Keywords: Post-transplantation lymphoproliferative disease, Ebstein-Barr Virus (EBV), Burkitt Lymphoma, Liver transplantation

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Screening for systemic cancer is an important part of both liver transplant recipient and donor preparation. During recipient preparation, if an extrahepatic malignancy is detected, an individualized treatment protocol should be established based on the type of malignancy and the severity of the patient's liver disease. Depending on the severity of the malignancy and the 5-year life expectancy, liver transplantation can be planned in some cases without a post-treatment waiting period, while in others it has been adopted by many transplant centers to remove the patient from the transplant list and perform transplantation after the follow-up periods recommended by the guidelines for malignancy treatment.[1] Institutional Review Board (IRB) approval was not required by IRB for the preparation of this report but written informed consent was obtained from the patient for publication.

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Non-Hodgkin lymphoma (NHL) accounts for about 90% of all lymphoma cases, with 85-90% arising from B lymphocytes and the remainder derived from natural killer (NK) and T lymphocytes.[1, 2] 25-35% of NHL cases arise from extra-nodular areas such as the small intestine, skin, and stomach. Secondary renal involvement may be observed in 30-60% of disseminated NHL cases; however, primary renal lymphoma (PRL) is rare and accounts for less than 1% of all renal masses.[3-5] PRL cases can be mistaken for RCC due to the peripheral localization of the mass and similar radiological findings. Early-stage RCC is a malignancy with a 5-year survival rate of over 95% when detected. However, due to different tumor biology and outcomes, pre-transplantation approaches vary between these two patient groups.[2]

In this report, we present an instance of LT for a patient incidentally diagnosed with high-grade PRL. Institutional Review Board approval was not required for the preparation of this report but written informed consent for publication was obtained from the patient.

Case Report

A 42-year-old diagnosed with history of Primary Sclerosing Cholangitis (PSC) was admitted to our center for LT for further decompensated liver cirrhosis. Child-Turcott-Pugh score was C10, and a MELD score was 19.

The physical examination revealed a poor general condition, widespread muscular atrophy, hepatosplenomegaly, jaundice, and Grade 2 hepatic encephalopathy. Preoperative radiological evaluation identified two lesions, measuring 23 mm and 8 mm, located on the lateral and medial sides of the upper pole of the left kidney, respectively, suggesting early-stage renal cell carcinoma (RCC). (Fig. 1) During the case-based Uro-Oncology meeting, it was agreed that removal of the masses would be appropriate following liver transplantation (LT) and normalization of liver functions, given the advanced liver disease, significant thrombocytopenia, and coagulation disorder.

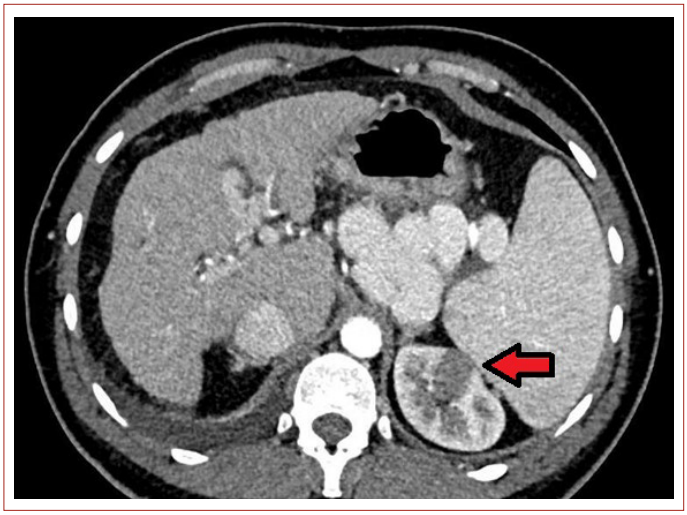

Figure 1. Left Kidney Upper Pole Lateral RCC, CT image.

The patient underwent a liver transplant of the right lobe and a splenectomy, featuring a single arterial anastomosis, a single portal vein anastomosis, and multiple hepatic vein anastomosis, receiving the organ from his sister who has the same blood type. To address the bile duct compromised by underlying PSC, a Roux-en-Y hepaticojejunostomy was performed for the biliary anastomosis.

On the postoperative day 5, despite medical treatment and endoscopic interventions, gastrointestinal bleeding persisted and entero-enterostomy revision was needed. Due to normalization of the liver functions and coagulation parameters, excision of renal masses was also performed. The patient was discharged on the postoperative 20th day uneventfully.

The pathological examination of the left-sided masses identified them as Burkitt lymphoma / High-grade B-cell lymphoma, not otherwise specified (NOS). The analysis showed B-cell neoplastic infiltration with necrotic changes, pronounced kappa light chain clonality, and high proliferative activity. Immunomarker assessment was revealed as following Leukocyte common antigen (LCA) (+), cluster of differentiation (CD) 79a (+), paired box protein (pax) 5 (+), CD19 (+), CD3 (-), CD20 (+), kappa light chain (+), Lambda Light Chain (-), CD10 (+), B-cell leukemia/lymphoma (BCL) 6 (+), BCL-2 (-), CD5 (-), cellular myelocytomatosis oncogene (CMYC) (-), Ki-67 95%

Hematology and oncology board suggested positron emission tomography (PET) scan and, no further lesion was detected. 4 cycles of adjuvant Rituximab, Cyclophosphamide, Doxorubicin and Prednisolone regimen were administered. The patient is followed up in the 2nd postoperative year of the procedure with normal liver functions and no recurrence.

The second-year follow-up evaluations of the patient after 4 cycles of adjuvant Rituximab, Cyclophosphamide, Doxorubicin and Prednisolone (R-CHOP) regimen was tumor free and liver functions were also normal.

Discussion

There is an increase in incidence of PRL which correlates with the increasing incidence of NHL related to development and availability of diagnostic modalities. There is a male predominance with being reported up to 1.6:1.[5] Most cases of PRL reported in the literature are unilateral, with bilateral renal involvement comprising less than 20% of the cases. The pathophysiology of PRL has been debatable as kidney is not a lymphatic organ and the origin of the lym-

phoma cells is not well clarified, and it is thought to be origin of the PRL is linked to lymph nodes of the renal sinus or the lymphatics of the renal capsule. Furthermore, PRL may be associated with history of chronic inflammation and pyelonephritis, Epstein-Barr Virus, and autoimmune diseases. [5, 6] On the other hand, extra-nodal NHL presenting as a solid lesion mimics other types of solid organ malignancies before histopathologic examination is performed, diagnosing PRL remains to be a challenge.[7]

Stallone et al. have proposed three diagnostic criteria for PRL: lymphomatous renal infiltration, no extra-nodal localization and non-obstructive unilateral/bilateral enlargement.[6] The diagnosis of PRL and its differentiation from other primary renal malignancies including RCC is critical especially in terms of further treatment. Ultrasonography may detect homogeneous, hypoechoic lesion in cases of PRL, however the imaging study of choice is computed tomography (CT) which demonstrates hypovascular masses with homogeneous attenuation unlike heterogeneity of RCC. Hypovascularity of PRL observed using color doppler also differs from the hypervascular pattern of RCC. [8] On CT scan, PRL typically presents with the following patterns: presence of multiple and bilateral hypovascular renal masses, presence of peri-renal disease, diffuse renal infiltration and renal invasion from contiguous retroperitoneal disease.[9] However, CT findings of PRL remain to be unspecific and histopathologic examination is considered gold standard in the diagnosis, with the most common histology type being the diffuse large B cell lymphoma (DLBCL) followed by marginal zone lymphoma. PET scan may be also useful in the diagnosis of PRL as NHL is intensely FDG-avid while RCC may not show intense FDG uptake.[10] Accurate diagnosis of PRL is vital as the survival and renal function has improved in PRL patients owing to the modern chemoradiotherapy, while RCC is managed surgically in most cases.[11] Histopathologic evaluation of PRL is also essential for grading which is the primary factor determining further management.

The use of CHOP (Cyclophosphamide, Doxorubicin, Vincristine and Prednisolone) has been reported for PRL in various case series, however it should be acknowledged that addition of rituximab could improve the survival.[12] The role of surgery in PRL is controversial, however, in more than 90% of case reports, nephrectomy has been performed with the differential diagnosis of RCC.[13] Due to being the most common subtype of PRL diffuse large B cell lymphoma, overall mortality of PRL despite new advances in treatment remains to be high. Delayed diagnosis also contributes to the poor survival. In their study of 599 primary renal lymphoma patients Taneja et al. report a median overall survival of 115 months. Overall male gender, stage III-IV, DLBCL histology and advanced age are considered as poor prognostic factors.

In connection with this information, in cases of lymphoma, it is recommended to wait 2 to 5 years after remission, as lymphoma relapse often occurs in this time frame.[14] In contrast, in patients with early stage RCC, there is no specific waiting period for liver transplantation. Depending on the severity of liver disease, even elective kidney surgery may be performed when liver functions return to normal after liver transplantation.[15]

In our patient, preoperative evaluations revealed two renal masses in the left kidney, which were initially thought to be consistent with renal cell carcinoma (RCC). The diagnosis was misled by the rarity of primary renal lymphoma and the more common occurrence of RCC. Fortunately, the masses were successfully removed via laparotomy, and an early diagnosis of "lymphoma" was established, leading to a good response to chemotherapy. However, had it been known that the renal mass was lymphoma, it would have been a contraindication for liver transplantation (LT) according to clinical guidelines, and the transplantation would not have been planned.

Furthermore, if preoperative biopsy or increased suspicion towards lymphoma were present, transplantation process would be reconsidered as defined by the guidelines with combined decision of transplantation team and haemato-oncology team. On the other hand, transplantation team should consider making a long-term transplantation plan for this specific type of patient group via close follow-up of their hematological progress whether remission is achieved without active disease in between 2-5 years. Individualized, risk-assessed and multi-disciplinary approach in patients with dire necessities for liver transplantation in this patient group is also a must to achieve both hematological and transplantation success. Risk stratification should be made more diligently in patients with more dire need of liver graft than need of hematological remission.

Cancer screening is one of the most important evaluations for liver transplant candidates. Especially in case of radiologic suspicion, tissue biopsy should be performed to establish the actual diagnosis. In addition, the severity of the patient's liver disease and the detected malignancy should be evaluated on a patient-by-patient basis, risk analysis should be performed, and the most appropriate treatment plan should be organized.

Disclosures

Informed consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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